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Clinical Study

# Medullary hemangioblastoma: 34 patients at a single institution

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ABSTRACT

This study aimed to elucidate the surgical experience of medullary hemangioblastoma (MH) at a single institution. We reviewed 34 consecutive patients with MH operated on between January 2005 and June 2012 in the neurosurgery department of the Beijing Tiantan Hospital. There were 14 men and 20 women. The patients were aged from 17 to 60 years with an average age of 38 years. Tumors were cystic in 12 patients (Type A), and solid in 22 patients. The solid tumors were of a small size in six patients (<3 cm, Type B), large in 12 (3.1–5 cm, Type C), and giant in four (>5 cm, Type D). Radical tumor removal was achieved in all patients. Tracheotomy was performed in 10 patients (one Type B patient, seven Type C, two Type D) postoperatively. Pneumonia secondary to lower cranial nerve palsy occurred in six patients (all Type C). Complications including intracranial infection (n = 5), gastrointestinal bleeding (n = 2), and intracranial hematoma (n = 1) also occurred in this group. Follow-up (range, 2–82 months; mean, 30 months) was available in all patients. At follow-up, 29 patients (85.3%) had a good outcome. Twenty-eight of these (82.4%) had an excellent outcome postoperatively (Karnofsky Performance Status $\geqslant$80). Although transient surgical complications are possible especially for large solid tumors, total surgical resection can be performed with favorable long-term outcomes with meticulous microsurgical technique and understanding of the vascular pattern of the tumor. Postoperative management of MH is as important as the operation.



## 1. Introduction

Intracranial hemangioblastomas account for 3.7% of all intracranial tumors at our hospital. These tumors are most commonly found in the cerebellar hemispheres (65%), followed by the vermis (15%), and cerebellopontine angle and brain stem (8%). The majority of hemangioblastomas are sporadic; about 20% are associated with von Hippel–Lindau (VHL) disease.[1–4] Although hemangioblastomas are histologically benign tumors, their operative management can be challenging, especially with brain stem involvement. To elucidate the impact of neurosurgical progress on the management of these particular tumors, we retrospectively reviewed our data from a consecutive series of 34 patients without VHL disease or multiple hemangioblastomas.

## 2. Patients and methods

### 2.1. Patient population

From January 2005 to June 2012 a total of 34 consecutive adult medullary hemangioblastoma (MH) patients underwent surgery by the senior author (L.Z.) in the neurosurgery department of the Beijing Tiantan Hospital, Capital Medical University, China. All 34 patients had a solitary MH and did not have a family history of hemangioblastoma or VHL disease.

All patients or their legal representative gave written informed consent to the study protocol, which was approved by the ethics committee of Beijing Tiantan Hospital.

### 2.2. Neuroimaging studies

All patients were evaluated pre-operatively with a CT scan and contrast-enhanced MRI. MRI is the diagnostic test of choice, as it clearly demonstrates the location of the tumor and its relationship to surrounding tissue.

We further divided the 34 patients with MH into four groups according to tumor size. Type A: predominantly cystic tumors (the tumor nodule varied from 0.1 cm to 1.5 cm); Type B: solid tumors of a small size (<3 cm); Type C: solid tumors of a large size (3.1–5 cm); and Type D: solid tumors of a giant size (>5 cm; Fig. 1).

### 2.3. Surgical procedure

All patients were operated on in the park-bench position. Electrophysiologic monitoring of the lower cranial nerves (LCN), somatosensory evoked potentials, and brainstem auditory evoked

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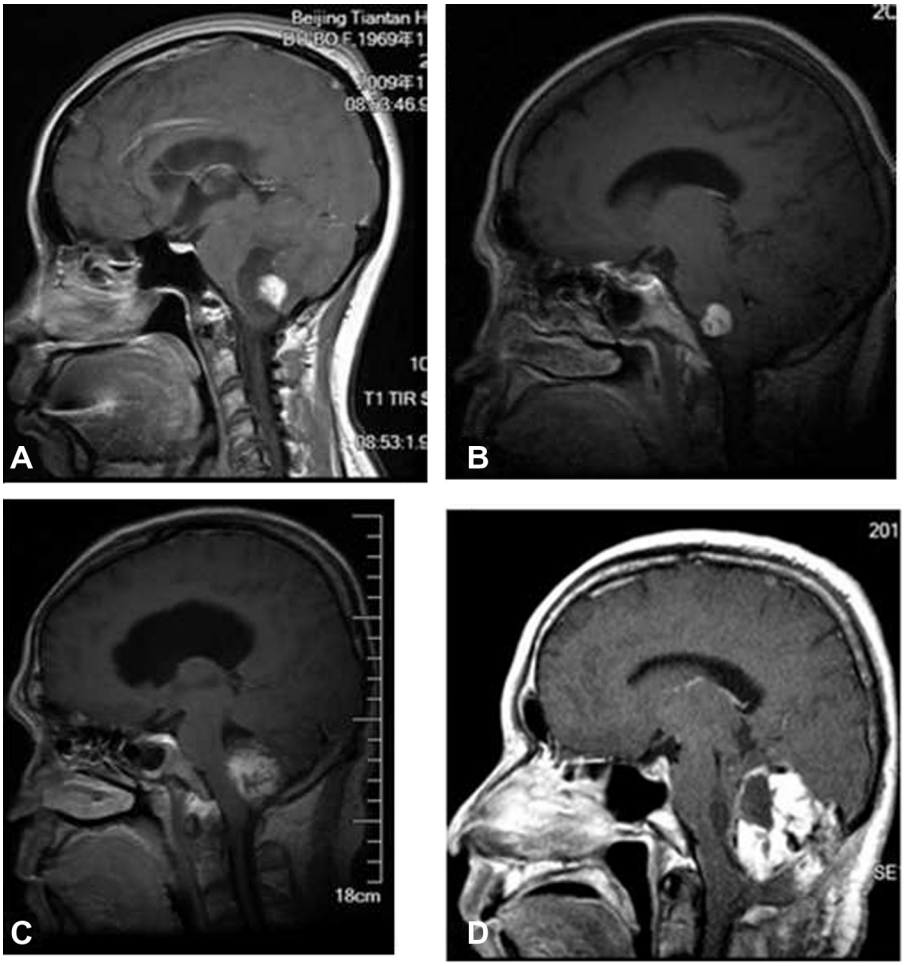


**Fig. 1.** Sagittal T1-weighted MRI with gadolinium contrast showing (A) a predominantly cystic tumor (Type A; tumor nodule 0.1–1.5 cm), (B) a small solid tumor (Type B; <3 cm), (C) a large solid tumor (Type C; 3.1–5 cm), and (D) a giant solid tumor (Type D; >5 cm). (This figure is available in colour at www.sciencedirect.com.)

potentials was used in all operations. The posterior midline approach was performed for posterior, posterolateral or intramedullary MH; thus, the lower part of the occipital bone and the posterior arch of the atlas were opened and enlarged on the side of the tumor.

For most anterior and anterolateral tumors, a far-lateral retrocondylar approach was chosen. An inverted hockeystick skin incision was used that was carried superiorly to the superior nuchal line, curving below the inion and proceeding down to the spinous process of C2.

### 2.4. Postoperative management

The postoperative care of patients depended on the operative procedure. Patients were sent to the neurosurgical intensive care unit (NICU) after surgery. Intubation was continued for at least 12 hours. If cough and gag reflexes were present on the second postoperative day, the endotracheal tube was removed. For patients who were at risk of aspiration pneumonia or dyspnea, a tracheotomy was performed to prevent respiratory complications. Nasogastric feeding provided nutritional support for dysphagic patients.

### 2.5. Follow-up

Follow-up data were available with clinical examination and MRI scans at 6 months and 1 year after surgery. Long-term follow-up in this study was based on our outpatient files and, for a few patients, a telephone call or home visit. Thereafter, patients were followed every 1 or 2 years.

## 3. Results

### 3.1. Clinical data

Fourteen patients were men and 20 were women (ratio 1:1.43). Their ages ranged from 17 to 60 years (mean, 38 years). The average duration of symptoms before presentation was 11 months (range, 0.5–120 months). Paresthesia was present in 16 patients (47.1%), headache in 15 (44.1%), LCN symptoms in 12 (35.3%), vomiting in nine (26.5%), cerebellar symptoms in eight (23.5%), dizziness in seven (20.6%), pyramidal signs in seven (20.6%) and neck pain in three (8.8%). Four patients (11.8%) in this series had undergone previous surgery, and one (2.9%) had received previous Gamma Knife radiosurgery (Elekta AB, Stockholm, Sweden). The pre-operative Karnofsky Performance Status (KPS) score was 74.7 ± 11.1 (mean ± standard deviation; Table 1).

### 3.2. Tumor characteristics and surgical aspects

According to the imaging appearance and that during operation, tumors were located in the medulla (n = 18), ponto-medulla(n = 13), and cervico-medulla (n = 3). Tumors were cystic in 12 patients (Type A), and solid in 22 patients. Tumors were small